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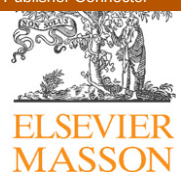

LETTER / *Cardiovascular imaging*

Endomyocardial fibrosis in the context of hypereosinophilic syndrome: The contribution of cardiac MRI

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MRI is a key tool for diagnosing, monitoring, and in the prognostic stratification of most cardiomyopathies [1]. It permits exhaustive study of systolic and diastolic function and precise tissue characterisation, particularly of fibrosis, using delayed-enhancement sequences (DE) [2,3]. We report here the case of a 79-year-old female patient of Caucasian origin referred for MRI to investigate disabling dyspnoea in the context of a hypereosinophilic syndrome.

Observation

This patient was referred to us for investigation of New York Heart Association (NYHA) grade 3 dyspnoea associated with a hypereosinophilic syndrome, with no other blood abnormality. The ECG showed incomplete right bundle branch block. Cardiac MRI was performed in this patient with cardiac gating, in regular sinus rhythm at 70 bpm. The left ventricular ejection fraction (LVEF) was subnormal at 50% with discrete global hypokinesis and a small left ventricle (LV) (normalised end diastolic volume = 63 mL/m^2, normal 41—81 mL/m^2). Right ventricular function (RV) was normal. First pass perfusion showed diffuse subendocardial hypoperfusion in the LV and RV (Fig. 1 a, b, c, arrows). DE sequences showed diffuse subendocardial enhancement involving both the LV and RV, without vascular systematisation, typical of endomyocardial fibrosis (Fig. 1 d, f, arrows). They also showed the presence of a thrombus at the apex of the LV and RV (Fig. 1 e, f, arrowheads). The atria were dilated with a 4-chamber view of the left atrium measuring 27 cm^2 at the end of systole (N < 24 cm^2).

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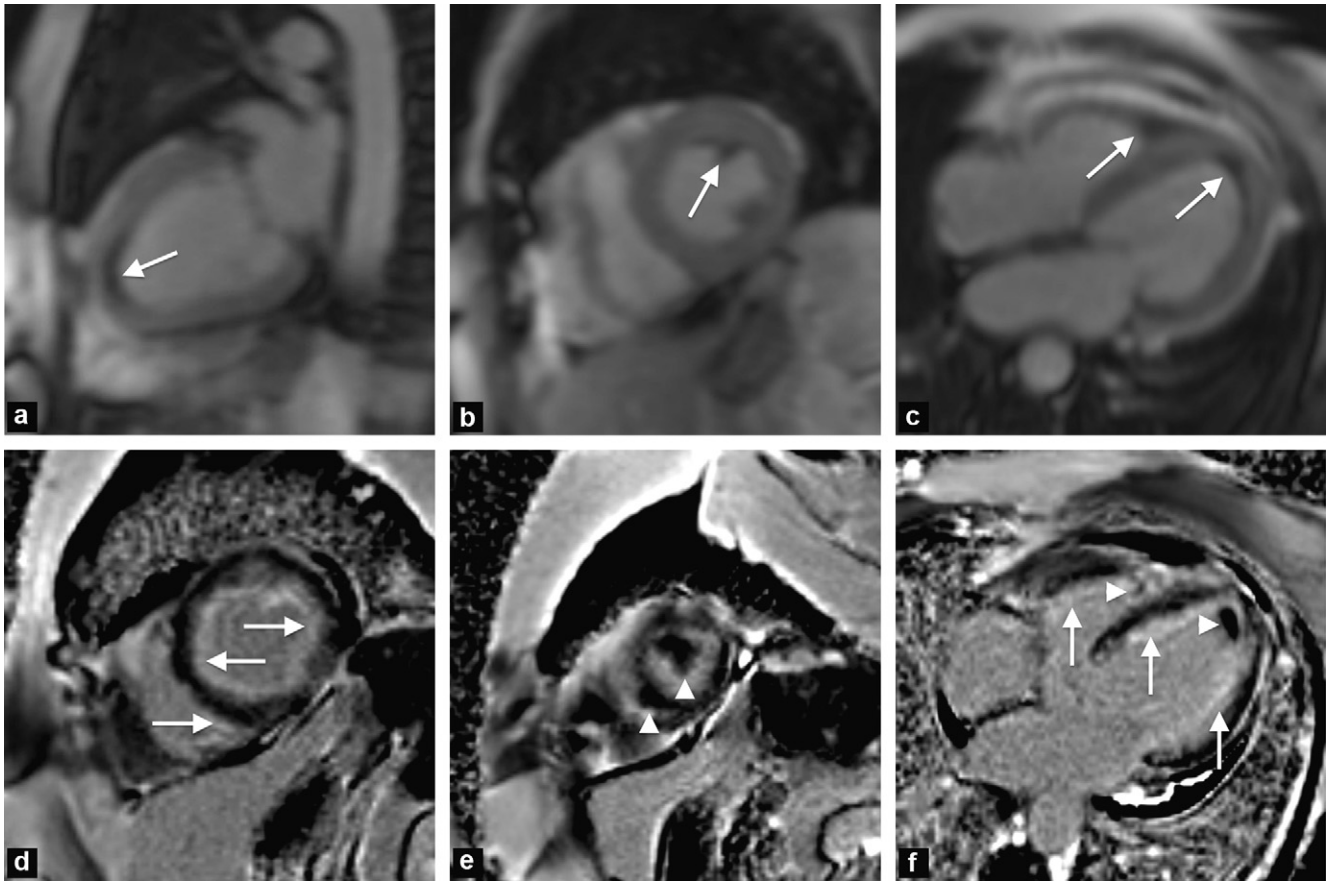

Figure 1. Cardiac MRI: first passage perfusion sequences (a, b, c) showing hypoperfusion as a hyposignal in the apices of the ventricles (arrows); late enhancement sequences (d, e, f) showing diffuse subendocardial enhancement (arrows) associated with the presence of thrombi in the apices of the ventricles (arrowheads).

Discussion

Endomyocardial fibrosis is a condition belonging to the group of restrictive cardiomyopathies first described in the 1940s in Uganda by Davies [4]. It is characterised by the deposition of fibrous tissue in the endocardium, predominantly in the filling routes and the apices of the right and left ventricles [5,6]. Although this is a condition that was originally described in tropical and subtropical regions, cases are also regularly described in Europe. Endomyocardial fibrosis lesions as described by Davies resemble the fibroplastic parietal endocarditis described by Löffler in temperate countries, which is found in idiopathic or secondary hypereosinophilic syndromes such as Churg-Strauss syndrome [7,8]. The term endomyocardial fibrosis is used in the literature to describe one or other of these entities. The lesions described in endomyocardial fibrosis also resemble those found in carcinoid heart disease [9]. This is because of the importance of the role played by eosinophils and serotonin in the pathological process. The mechanisms of this disease are still debated and other causes have been suggested: infections (toxoplasmosis, rheumatoid arthritis, malaria, helminthiasis), allergies (hypereosinophilia, autoimmunity), malnutrition, and toxic agents. In practice, transthoracic echocardiography is the first examination to be undertaken and provides an accurate assessment of systolic and diastolic function. Nevertheless, it has limitations for characterisation of tissues and differential diagnosis of restrictive cardiomyopathies. MRI, however, plays a key role in the diagnosis and prognosis of this condition, although few data have been reported in the literature [10—12]. It provides precise morphological evaluation, usually characterised by restriction with non-dilated or small ventricles, as in the case reported here. The atria are often increased in size because of the severe diastolic dysfunction with a restrictive disorder [2]. This diastolic dysfunction is the cause of the patients' symptoms who often have disabling NYHA grade 3 or 4 dyspnoea. The DE sequences confirm the diagnosis by showing typical DE, limited to the subendocardium and extending from the subvalvular regions to the apices of the two ventricles, where it usually predominates. A key element is that the enhancement is not distributed in the vessels and, in most cases, is not accompanied by myocardial thinning. A thrombus is frequently present at the apex of the LV and/or RV and, here again, MRI plays a key role in providing this diagnosis, which is often underestimated by echocardiography. Recently, a prognostic role for MRI has also been suggested [11]. The standard treatment, indicated in patients with NYHA grades 3 or 4, is surgical resection of areas of endomyocardial fibrosis [5,6]. MRI may in the future help in planning surgery and monitoring its efficacy. In the case of our patient, it was difficult to envisage surgical treatment given her extremely fragile condition. Medical treatment combining diuretics and converting enzyme inhibitors improved the dyspnoea.

Follow-up MRI after one year (not shown) showed stable functional abnormalities and delayed enhancement. It also showed marked regression of the apical thrombi under anticoagulant treatment.

Conclusion

Endomyocardial fibrosis is a rare disease in temperate countries, and falls within the differential diagnosis of restrictive cardiomyopathies. Cardiac MRI provides a precise aetiological diagnosis for patients referred with suspected endomyocardial fibrosis, and could help prognostic stratification and in planning therapeutic procedures for these patients.

Disclosure of interest

The authors declare that they have no conflicts of interest concerning this article.

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